A Huge Congenital Left Atrial Appendage Aneurysm

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Left atrial appendage aneurysm (LAAA) is an extremely rare cardiac anomaly which is mainly characterized by localized or diffuse dilatation of left atrial appendage.[1] This condition with diverse symptoms including arrhythmia, thromboembolic events, and heart failure confuses physician and is easily misdiagnosed.[2] We herein report a case of huge LAAA which is misdiagnosed originally.

A 33-year-old female patient presented to us with intermittent chest discomfort for 2 years and sudden syncope 1 month before without coronary heart disease, hypertension, or valvular heart disease. She was diagnosed as pericardial cyst by a physician of local hospital with transthoracic echocardiography (TTE), and her thoracoscopic resection was discontinued when the surgeon observed the left atrial enlargement during operation. After transferring to our hospital, the condition was reassessed with stable vital signs, normal physical examination, and sinus rhythm by electrocardiography. Chest X-ray showed cardiac enlargement and prominent left atrial appendage. TTE [Figure 1a] showed a cystic intrapericardial free-echo formation of 8.4 cm × 6.8 cm size communicating with left atrium and compression of left ventricular with normal ejection fraction of 67%. Cardiac computed tomography (CT) scan [Figure 1b] conformed TTE results and showed a huge LAAA connected to the left atrium through a 23 mm neck.

Considering the history of syncope and compression of left ventricular, we indicated surgical treatment with aneurysmectomy of LAAA through median sternotomy and cardiopulmonary bypass. The neck of LAAA was ligatured with silk, and no thrombus was observed after incision of aneurysm wall. The aneurysm was resected entirely with a 4-0 Prolene polypropylene continuous suture. Intraoperative TEE confirmed no residual atrial aneurysm after resection. Pathological examination confirmed atrial aneurysm which consisting of myocardial tissue with fat infiltration.

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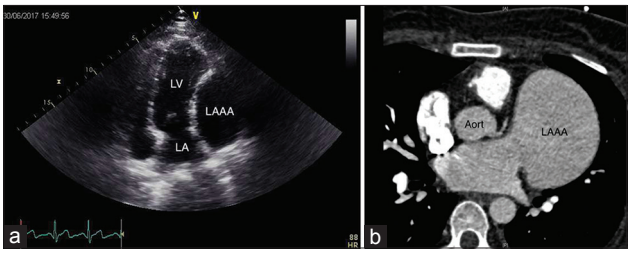

Figure 1: (a) Transthoracic echocardiography showing a giant LAAA compressing the LV. (b) Cardiac computed tomography showing the relationship between the appendage aneurysm and the left heart. LAAA: Left atrial appendage aneurysm; LV: Left ventricular.

The postoperative course was uneventful, and we noticed disappearance of chest discomfort and syncope. Postoperative TTE showed no evidence of LAAA and the patient discharged in good clinical condition with a regimen of metoprolol tartrate 12.5 mg daily. At 2-week follow-up, she reminded asymptomatic and continued to be sinus rhythm with no arrhythmia.

LAAA has been classified as intrapericardial or extrapericardial according to integrality of pericardium and can also been classified as congenital or acquired, two-fifths of which are congenital and the rest occur due to surgery, trauma, and valve disease (mitral stenosis and/or mitral regurgitation).[3] The cause of LAAA is still unclear, and growth in congenital cases may result from depauperation of dysplastic pectinate muscles, which changes the LAA from a pump to a reservoir and leads to progressive dilation resulting

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in elevated internal pressure. LAAA has been reported to occur concomitant with other congenital anomalies such as atrial septal defect, ventricular septal defect, and anomalous renal artery in the literature. LAAA has been reported across all the ages but most common in the third decade.[3]

Symptoms usually do not arise with small aneurysms. Once they reach a larger size as patients' ages increase, palpitations and/or dyspnea are the most common symptoms, owing to triggered or reentrant atrial tachyarrhythmia in the enlarged left atrium, abnormalities of the conduction system, and compression of cardiovascular or airway.[4] The life-threatening complication is thrombosis due to stasis of blood in LAAA which can lead to systemic embolism once shedding off, most common in brain and limb. Chest X-ray usually shows cardiomegaly with a prominent left atrial appendage, but not specific. Electrocardiogram and 24-h Holter monitoring may reveal supraventricular arrhythmia. TEE is considered a primary method to identify LAAA and other cardiac abnormalities but with a limited sensitivity of 45%.[5] In our case, the patient was mistaken for pericardial cyst in her initial diagnosis by TEE since no aneurysm neck was identified and suffered wrong operation. Transesophageal echocardiography (TEE) is superior to TTE for the detection of LAAA with a sensitivity of 90% which can provide more detail and clear visualization for aneurysm neck, blood flow, and tiny thrombus. Cardiac CT or magnetic resonance may be used as an adjunct for better definition of anatomy, studying the relationship of LAAA with neighboring structures and confirming alternate diagnoses and associated congenital anomalies.

To treat our patient validly, 94 LAAA case reports published from 1980 to 2017 were reviewed using "left atrial appendage," "left atrial appendage aneurysm," and "left atrial aneurysm" by searching MEDLINE. Eighty patients (85.1%) underwent surgical treatment regardless of patient's symptoms or the size of aneurysm. Recommended surgical strategy is the use of cardiopulmonary bypass through median sternotomy including aneurysmectomy in 49 cases (61.3%) and excision of the left atrial appendage in 23 cases (28.8%). Endoscopic resection or stapling of aneurysm with off-pump is used in the rest cases which may increase surgery risks. Ten cases (10%) went concomitant ablation procedures for their atrial fibrillation. Most patients had no reports of advent events during follow-up except for 3 patients (3.8%) who had postoperative thromboembolic adverse events and 2 patients (2.5%) experienced cardiac arrhythmia. Medical treatment, including beta-blockers and anticoagulants, are empirical strategy with limited literature and no randomized trials. Our patient was indicated to resection of huge LAAA with typical symptoms for dyspnea and syncope, which showed satisfying clinical outcome.

In spite of a rare disease, LAAA is recommended to early intervention to prevent thrombotic events, severe arrhythmia, and cardiac function deterioration even in asymptomatic cases because the risks of operation are relatively low and outcomes are generally satisfying.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient has given her consent for her images and other clinical information to be reported in the journal. The patient understands that her name and initial will not be published and efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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